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Solitary fibrous tumor of the anterior abdominal wall. A case report and review of the literature

BACKGROUND: *Solitary fibrous tumor (SFT) is a rare mesenchymal neoplasm affecting soft tissues with a not well defined biological behavior. SFT occurs mostly in the pleura and the thorax, while extra-thoracic localization is uncommon and abdominal localization is very rare. Histologically, SFT is a well defined mass with splindle-cell proliferation in collagenous matrix with staghorn vascular network and CD34 reactive.*
CASE REPORT: *A 64 years-old man with a history of recurrent gastric cancer previously treated with total gastrectomy, was admitted with contrast enhanced CT-scan diagnosis of a well demarcated oval mass of 4.8 cm with microcysts, vascularized in the arterial phase and with wash out in the tardive phase, located in the peritoneal side of right rectus abdominis muscle, suspected for metastatic gastric tumor. The patient underwent minilaparotomy and en-bloc excision of the lesion. Histologically the tumor was characterized by a hemangiopericitoma like growth pattern and the immunostaining was positive to CD34, CD99, BCL-2 and Vimentin. The definitive diagnosis was SFT with a proliferation index (Ki-67/MIB-1) <3%. In our case, chemotherapy was not indicated. At the 6-month follow-up, the patient is in good clinical conditions with no recurrence or metastasis.*
CONCLUSIONS: *We reported a rare case of primitive SFT located in peritoneal side of the of right rectus abdominis muscle treated surgically, in a patient previously affected by gastric adenocarcinoma. In this case, SFT showed a benign behaviour during a short term follow-up. Dimensional pattern, histopathological features and curative surgery remain the most important indicators of clinical outcome.*

KEY WORDS: Abdominal wall, Hemangiopericitoma, SFT, Solitary fibrous tumorSpindle cell.

Introduction

Solitary fibrous tumors (SFT) is a rare mesenchymal neoplasm affecting soft tissues, often localized in the thoracic cavity, mostly in the pleura [1].
Extra-thoracic localization is unfrequent and usually occurs in the meninges and lower extremities; abdominal or pelvic localization is rare [1]. SFTs represent less than 2% of all soft-tissue tumors and commonly occur in middle-aged adult [2]. In most cases SFTs are benign lesions, with malignant behavior described in 10 to 36% of cases [3]. The 2002 World Health Organization scheme classifies SFTs as intermediate biological potential (rarely metastasizing) [4]. Dimensional criteria and local invasion are important to predict the probability of recurrence or metastasis since it has been observed that SFTs greater than 10 cm have worse outcome and deserve a closer follow-up [5].
We present an unusual case of SFT of the anterior abdominal wall located in the peritoneal side of the rectus abdominis muscle with the revision of the current literature.

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Case Report

A 64 years old man with a history of recurrent gastric cancer treated with total gastrectomy, was admitted to our department with ultrasonographic and contrast enhanced CT-scan incidental findings of a well demarcated oval mass of 4.8cm with microcysts, vascularized in the arterial phase and with wash out in the tardive phases (Figs. 1,2). The mass was located in the peritoneal side of right rectus abdominis muscle, in the context of mesenteric adipose tissue, without continuity with other organs or locoregional lymphadenopathy. The patient was asymptomatic. Laboratory findings were normal, including tumor markers CA 19.9, CEA, PSA. The patient underwent minilaparotomy with an en-bloc excision of the mass including the peritoneum layer and the muscolaris fascia. Macroscopically, the solid mass appeared partially cystic, with a grey cut-surface and its dimension was 4.2x3x2.5 cm. Microscopically the tumor was characterized by spindle cell proliferation with poor cytoplasm and indistinct outlines (Fig. 3). These cells had vescicular nucleus and were separated by collagen bundles showing a hemangiopericitoma like growth pattern (Fig. 4). The immunostaining was positive to CD34, CD99, BCL2 and Vimentin (Figs. 5,6). The definitive diagnosis was SFT with a proliferation index (Ki-67/MIB-1) <3%. The post operative period was uneventful and the patient was discharged in 2nd post-operatory day in good clinical conditions. In our case, chemotherapy was not indicated. At the 6-month follow-up, the patient is in good clinical conditions with no recurrence or metastasis.

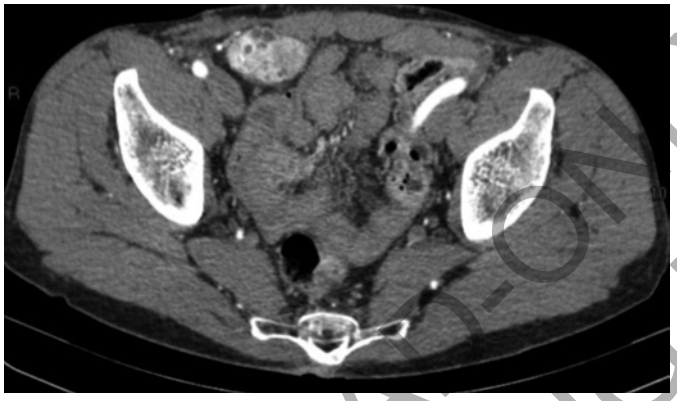

Fig. 1: CT scan images. Contrast enhanced CT-scan showed a well demarcated oval mass with microcysts vascularized in the arterial phase.

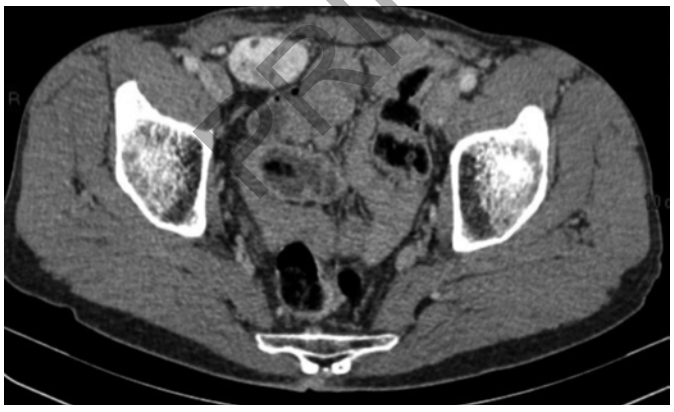

Fig. 2: CT scan images. Contrast enhanced CT-scan showed a well demarcated oval mass with microcysts with wash-out in the portal tardive phase.

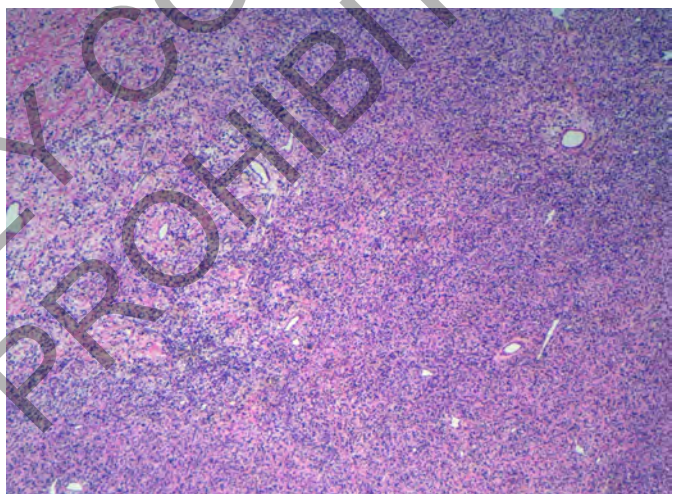

Fig. 3: Microscopics observations. The tumor shows typical spindle cell pattern. E.E. 4x.

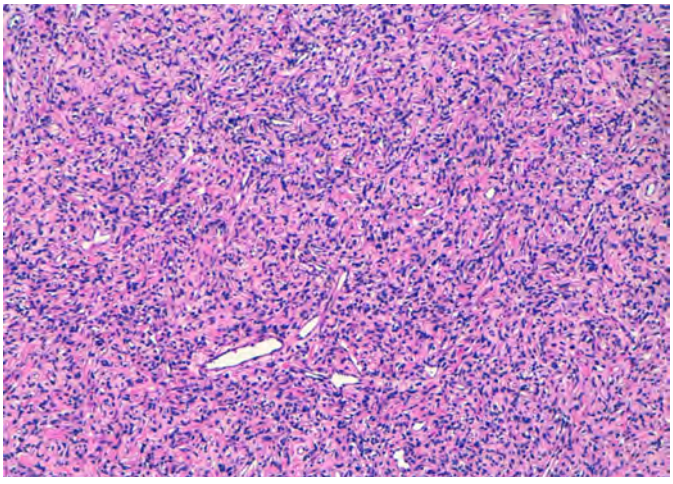

Fig. 4: Microscopics observations. The tumor shows Hypercellular area in collagenous matrix and hemangiopericitoma like vascular network. E.E.10x

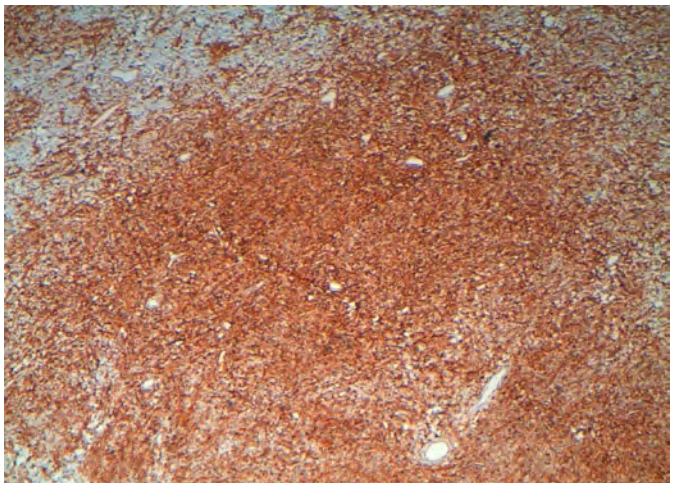

Fig. 5: Immunohistochemical examinations. The cells showed diffuse positivity for CD99. IHC 2.5x.

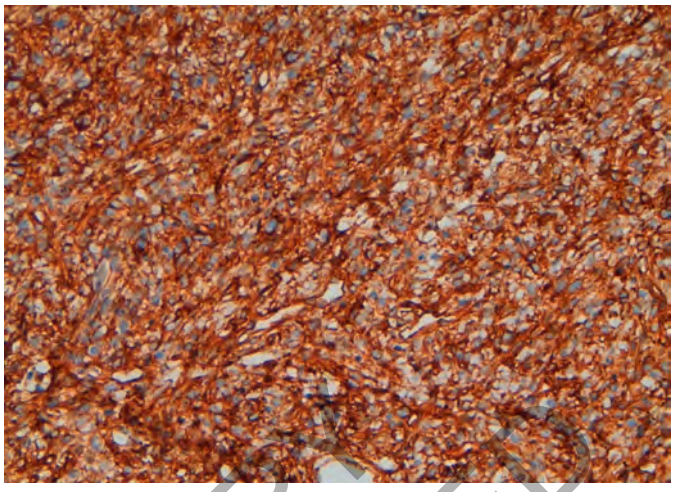

Fig. 6: Immunohistochemical examinations. The cells showed diffuse positivity for CD34. IHC 20x.

Discussion

SFT represents a single spectrum of mesenchymal tumors, of which hemangiopericytoma is a cellular phenotypic variant. SFT was first described in 1931 as a distinctive pleural lesion [6]. SFTs usually arise in the pleura [7], but they can occur at any site of the body in somatic soft tissue [8]. Classically, SFTs are composed of variably pleomorphic spindle cells admixed with collagen and arranged haphazardly or in short fascicles 4[6]. Alampady et al. have reported 13 cases of abdominal wall SFTs. SFT of the peritoneum occurs in both genders and in all generations, but predominantly affects those between the 4th and the 7th decades of life, with the benign variant of the tumor being three to four times more common than the malignant [9]. Preoperative diagnosis of SFT is difficult because clinical symptoms, physical examination and imaging findings using US, CT and MRI are not indicative 9[6]. Most SFT of deep soft tissue are indolent and are incidentally found during examinations for other disorders [10]. Clinically, extrapleural lesions present with symptoms related to the tumor site, sometimes resulting in compression symptoms. Rarely it may cause systemic symptoms such as hypoglycemia, arhralgia, osteoarthritis and clubbing. These symptoms usually resolve upon removal of the tumor [11,12]. Extrapleural SFT has been described as well-defined mass with a solid and cystic degeneration or hemorrhage [11,13,14]. Fibrosarcoma, malignant fibrous histiocytoma, leiomyosarcomas, hemangiopericytoma, synovial sarcoma and malignant mesenchymomas should be included in the main differential diagnosis. A correct final diagnosis requires pathological and immunohistochemical examination [9]. SFT shows a wide spectrum of histological features including the cellular, fibrous, fat-forming and giant cell-rich variants [12]. Usually, SFTs are histologically characterized by admixed hypocellular and hypercellular foci of proliferating spindle cells surrounded by collagenous stroma with hemangiopericitoma like appearance and prominent vascularity [7,10,15,16]. Immunohistochemically, SFTs commonly express CD34 and CD99. Bcl-2, EMA and SMA may be expressed, and they are usually negative for S100 protein, desmin and cytokeratins [12].To date, no definitive markers have been made to define a risk assessment model based on patient age, tumor size and mitotic index with promising results [17]. The most effective therapy in patients with SFT is oncological resection of the tumor [9]. The quality of the initial excision, with free margins, represents important prognostic factors [12]. During the follow-up, CT scan or MRI is necessary at least once a year since recurrence of SFT as long as seven years after resection has been documented [6,9]. Therapeutic options for more advanced or aggressive tumors are scarce and no standard modality for metastasized SFT has currently been accepted [17]. Few chemotherapeutical drugs have been studied with different outcome. Anti-angiogenic therapy such as monoclonal antibodies against growth factors shows interesting results for the treatment of unresectable SFT [17-19]. The majority of SFTs are benign, but those lesions may occasionally give rise to metastatic disease in the absence of any predictive morphological features. Histology-based characterization is frequently discordant with the clinical behavior of SFT and the outcome of patients with SFT is unpredictable based on the histological assessment [10]. For this reason SFT has been classified into the intermediate (rarely metastasizing) category [8]. The definition of malignancy involves the degree of local invasion, local recurrence and distant metastases. The most important prognostic factor in pleural SFTs is not the histological appearance but the resectability of the tumor [9,20]. Infact, pleomorphism, mitotic figures and necrosis do not necessarily predict the aggressiveness of the tumor; instead, infiltrative features that prevent total

respectability are significant for the outcome [10,16]. Recent studies showed no difference between intra and extra-thoracic SFT with regards to rates of malignant pathological diagnosis, with extra-thoracic SFT having a small but significant higher rate of locoregional recurrence suggesting more aggressive clinical behavior [3,5]. It is estimated that the rates of recurrence, intra-thoracic and extra-thoracic metastases for intrathoracic and extrathoracic SFT are similar (respectively 9-19%, 0-36%, 0-19%), with common sites of metastasis including the lung and liver (18). It has been observed that tumor with a diameter >10 cm have worse outcome [5]. Ten-year survival rate was reported to be between 54% and 89% after complete surgical resection with clear margins [21]. Careful long-term follow-up is therefore recommended for patients with SFT diagnosis[6].

Conclusions

We reported a rare case of primitive SFT located in the peritoneal side of the right rectus abdominis muscle treated surgically, mimicking locoregional solid metastasis at the first preoperative diagnosis, in a patient previously affected by gastric adenocarcinoma. In this case, SFT showed a benign behaviour during a short term follow-up. Dimensional pattern, histopathological features and curative surgery remain the most important indicators of clinical

Riassunto

Il tumore fibroso solitario (SFT) è un tumore mesenchimale raro che colpisce I tessuti molli. Tale tumore non ha un comportamento biologico ben definito e si localizza prevalentemente a livello pleurico e toracico. La localizzazione extra-toracica è rara e quella addominale è ancora più infrequente. Morfologicamente, il tumore fibroso solitario si presenta come una massa solida ben definita con proliferazione a cellule fusate in matrice collagenosa, vascolarizzazione "staghorn-like" e positività al CD34.

Riportiamo il caso clinico di un paziente di 64 anni con una storia di pregresso cancro gastrico trattato con una gastrectomia totale che si presentava alla nostra osservazione per il riscontro alla TC con mdc di follow-up, di una massa ovale con margini ben definiti di circa 4.8 cm di diametro localizzata sul versante peritoneale del muscolo retto dell'addome di destra. La massa si presentava microcistica al suo interno e vascolarizzata nella fase arteriosa, con wash-out nella fase tardiva. Il paziente non presentava sintomi e il quadro radiologico non permetteva una diagnosi differenziale tra metastasi da cancro gastrico o altra patologia neoplastica. Gli esami di laboratorio si presentavano nella norma. Il paziente è stato sottoposto ad intervento chirurgico con minilaparotomia e resezione en-bloc della lesione. All'esame istologico post-operatorio, il tumore presentava un pattern di crescita tipo "hemangiopericitoma-like" e positività per CD34, CD99, BCL-2 e Vimentina con diagnosi finale di tumore fibroso solitario e un indice di proliferazione (Ki-67/MIB-1) inferiore al 3%. Nel nostro caso, la chemioterapia post-operatoria non risultava indicata. A 6 mesi dall'intervento il paziente si presenta in buone condizioni cliniche generali con nessun segno di recidiva o metastasi alle indagini di follow-up. Dato il comportamento biologico incerto del tumore, si raccomanda comunque un follow-up a lungo termine. I criteri dimensionali, le caratteristiche istopatologiche e di proliferazione e la radicalità della chirurgia rappresentano gli indicatori più importanti di outcome clinico.

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